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Abstract: The authors present a microsurgical technique for the resection of a heterogeneous group of pineal-region tumors and discuss the key points for successfully performing this surgery. Twenty-six consecutive patients with pineal-region tumors were resected by the senior author (H.B.) and analyzed retrospectively. For all 26 patients, the operation was conducted using the infratentorial supracerebellar (ITSC) approach in the sitting (23 patients) or Concorde (three patients) positions. Twenty-five patients had symptomatic obstructive hydrocephalus and were treated with ventricular drainage, a previously inserted ventriculoperitoneal shunt, or an endoscopic third ventriculostomy before undergoing resection of the pineal-region tumor. The gross total removal of the tumor was achieved in 23 patients and subtotal removal was achieved in three patients. The tumors were pathologically diagnosed mainly as pineocytomas (10), pilocytic astrocytomas (6), or pineal cysts (4). Twenty-five of the patients clinically improved after surgery, and there was no mortality. Two patients experienced transient postoperative neurological deterioration: one patient developed Parinaud syndrome, and one patient developed intermittent diplopia. Successful surgery and patient outcome when treating tumors of the pineal region using the ITSC approach requires: (i) preservation of the venous flow of the Galenic draining system; (ii) preservation of the thick bridging veins of the tentorial surface of the cerebellum, especially the hemispheric bridging veins; and (iii) minimizing retraction of the cerebellum during surgery to avoid adverse effects caused by both direct cerebellar compression and disturbance of the venous circulation.

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Neurosurgical venous considerations for tumors of the pineal region resected using the infratentorial supracerebellar approach.

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Keywords

brain retraction, bridging vein, Galenic draining group, infratentorial supracerebellar approach, pineal region tumor

Abstract

Background: The authors present a microsurgical technique for the resection of a heterogeneous group of pineal-region tumors and discuss the key points for successfully performing this surgery.

Methods: Twenty-six consecutive cases of pineal-region tumors were resected by the senior author and analyzed retrospectively. For all 26 patients, the operation was conducted using the infratentorial supracerebellar (ITSC) approach in the sitting (23) or Concorde (3) positions.

Results: Twenty-five patients had symptomatic obstructive hydrocephalus and received ventricular drainage, a previously applied ventriculo-peritoneal shunt, or an endoscopic third ventriculostomy before undergoing resection of the pineal-region tumors. The gross total removal of the tumor was achieved in 23 patients and subtotal removal was achieved in three patients. The tumors mainly were pathologically diagnosed as pineocytomas (10), pilocytic astrocytomas (6), or pineal cysts (4). Twenty-five of the patients clinically improved after surgery, and there was no mortality. Two patients experienced transient postoperative neurological deteriorations: one patient exhibited Parinaud syndrome, and one patient presented intermittent diplopia.

Conclusion: Achieving a successful operation and patient outcome when treating tumors of the pineal region using the ITSC approach requires i) preserving venous flow of the Galenic draining group, ii) preserving the thick bridging veins of the tentorial surface of the cerebellum, especially the hemispheric bridging veins, considering the collateral circulations of the bridging veins, and iii) minimizing retraction of the cerebellum during surgery to avoid adverse effects caused by both direct cerebellar compression and the disturbance of venous circulation.

Introduction and Overview

Neurosurgical resection of tumors located in the pineal region is indicated, especially for benign tumors, excluding germinomas (1-5). However, as the lesions are deep-seated and located near important neural structures and bind to or associate with important veins of the Galenic venous draining group, surgery on pineal-region tumors is still challenging for neurosurgeons. In this report, we summarize and discuss our neurosurgical strategies and preoperative planning for this group of tumors and report our ten-year experiences in the microsurgical resection of pineal-region tumors via the infratentorial supracerebellar (ITSC) approach. The cases presented here focus on the microsurgical techniques and key considerations regarding the venous surroundings that enable successful surgery on the pineal-region tumors.

<u>Neurosurgical indication and timing of surgery</u>

Neurosurgical resection is indicated in patients showing one or more clinical symptoms because of the location of the pineal region and to ensure a histopathological diagnosis (1). Symptomatic benign tumors or cystic lesions can be cured by surgical resection alone (2). For malignant parenchymal or glial tumors, mass reduction is also valuable before starting radiation and chemotherapy (6, 7). In the case of the intraoperative diagnosis of germinomas during instantaneous section, further resection is unnecessary due to its high sensitivity to radiation and/or chemotherapy (7, 8). If a malignant germ-cell tumor is strongly suspected preoperatively, due to elevated concentrations of α-fetoprotein (AFP) and/or β-human chorionic gonadotropin (β-

HCG) measured by preoperative examination of serum or cerebrospinal fluid (CSF) (1, 9), chemotherapy combined with radiation therapy may precede the surgery, and neurosurgical resection should only be planned for residual tumors (7). If the patient's symptoms are caused by obstructive hydrocephalus, its treatment should precede the tumor resection. Temporary ventricular drainage, a permanent ventriculo-peritoneal (VP) shunt, or an endoscopic third ventriculostomy are possible treatments for obstructive hydrocephalus (9). Surgical resection of the tumor should then be performed.

Radiological imaging

Contrast-enhanced magnetic resonance (MR) images and multi-detector computed tomography (CT) provide useful information about tumor location, direction of extension, and relationship with surrounding structures (6). The importance of the venous phase of conventional cerebral angiography was traditionally emphasized. MR venography, CT angiography, and rotation digital-subtraction angiography also provide surgeons detailed, three-dimensional information about veins belonging to the Galenic draining group, bridging veins, and straight sinus. Surgeons must use those radiological findings to predict the individual morphological features of each patient before each surgery.

Selection of surgical approach

The choice of approach is guided by tumor size, main location of the tumor mass, direction of the tumor growth, relationship between the tumor and its surrounding structures, angle of the straight sinus, and surgeon's experiences (6). The infratentorial supracerebellar (ITSC) approach (10-12) and the occipital transtentorial (OTT) approach (13-15) are the standard approaches for the microsurgical resection of pineal-region tumors. The ITSC approach is a midline approach and provides definite orientation of the tumor and surrounding structures (16). It also allows a direct view of the tumor without interposition of veins of the Galenic draining group and facilitates the dissection of the tumor from its attachments along the velum interpositum and deep venous system (1, 15, 17). A disadvantage of this approach is the narrow angle to the lateral and caudal side of the surgical field (3, 4, 18) and the restriction of the operative field if the tentorium is inclined steeply to the axial plane (19).

In contrast, the OTT approach provides wider exposure than does the ITSC approach and is often preferable for tumors extending superiorly, caudally, and laterally (6, 20). This approach can be undertaken in various patient positions, such as lateral, prone, or combined positions, compared with the ITSC approach, in which a sitting or Concorde position is most appropriate. However, the OTT approach has the disadvantage of proceeding through the deep venous system that overlays the tumor, forcing the surgeon to work around the system to avoid injuries to the veins (16, 20, 21). The OTT approach also presents the risk of leading to occipital lobe injuries, followed by defects of the patient's visual field, due to the necessity of retracting the occipital lobe or sacrificing occipital bridging veins. As it is not a midline approach to the tumor, it is difficult to employ the OTT approach to evaluate the contralateral quadrigeminal region and the ipsilateral pulvinar in the posterior third ventricle (6, 18).

Selection of patient positioning

The sitting position is used most commonly for the ITSC approach (4, 11) because gravity minimizes the necessity for retraction of the cerebellum, facilitates the dissection of adherent veins from the tumor, and decreases the bleeding from the tumor by reducing the pooling of venous blood. However, use of the sitting position may cause critical systemic complications, such as air embolism and hypotension. These complications must be prevented by careful surgical hemostasis and through monitoring by neuroanesthesiologists.

To avoid the potential complications caused by the sitting position, the Concorde position was introduced for the ITSC approach (11). This position combines elements of the prone and sitting positions and gives surgeons access to the pineal region that is as good and straight as that provided by the sitting position, as well as reducing possible air embolisms. In the past, we preferred the sitting position in operations employing the ITSC approach, but sometimes used the Concorde position early in the series of operations reported here for patients with high-risk profiles, patients more than 70 years old, or children. Most recently, the Concorde position has been favored by our group for patients of any age.

Methods

We performed a retrospective analysis of patients surgically treated for pineal-region tumors in our department from August 1997 through July 2006. Twenty-six operations on 15 male and 11 female patients with a mean age of 31.6 years (nine to 61 years old) were performed by the senior author and were analyzed retrospectively. In all cases, the ITSC approach was used. The anatomical cadaver study of the pineal region was performed at the Neuroanatomical Dissection Laboratory in Zurich by the first author.

Results and Technical Considerations

Anatomy of the pineal region for the ITSC approach

The pineal gland protrudes from the posterior wall of the third ventricle and is deeply seated in the quadrigeminal cistern. The pineal region is defined dorsally by the splenium of the corpus callosum and tela choroidea, ventrally by the quadrigeminal plate, rostrally by the posterior third ventricle, caudally by the vermis of cerebellum, and laterally by the thalami and the tentorial edges (Figure 1).

The pineal region contains many veins draining into the vein of Galen (Figure 1) (22-24). The vein of Galen drains into the straight sinus beyond the pineal region. Most of these veins must be completely preserved during removal of the tumor to avoid serious surgical complications, such as thalamic venous infarction. If the tumor is large and a wide surgical space is necessary to remove it, only the precentral cerebellar vein (or the superior cerebellar vein) can be sacrificed among the veins of the Galenic draining group.

The pineal body is supplied by one or more pineal arteries, which are the branches of the medial posterior choroidal arteries (Figure 1). The P3 segments of the posterior cerebral arteries also run around the lateral parts of the pineal region.

When surgeons approach the pineal region using the ITSC route, they find several bridging veins between the cerebellar surface and the tentorium (Figure 3A, 4A). These bridging veins are divided into two groups (17, 25, 26). Those belonging to the vermian group receive venous blood from the inferior vermian veins and other superficial veins and drain to the torcular Herophili. The veins belonging to the hemispheric group receive venous blood from the superior and inferior hemispheric veins and drain into the tentorial sinus. As there are frequent communications among these veins, some of the bridging veins can be sacrificed without adverse effects. However, if bridging veins of the vermian group must be sacrificed to provide sufficient space in the pineal region, those of the hemispheric group should be preserved to maintain collateral venous circulations and prevent complications.

<u>Microsurgical ITSC technique</u>

The ITSC approach begins with a midline linear skin incision 4 cm above the inion to the spinous process of the axis (Figure 2). Bilateral suboccipital craniotomy follows with its upper line placed above the superior margin of the transverse sinus. The dura mater is incised transversely 5 to 10 mm below the inferior margin of the transverse sinus after the occipital sinus is ligated and divided (Figure 2). The superior edge of the dura mater is drawn upward by dural tenting sutures to gain a wider surgical view and to protect the transverse sinus.

Arachnoid adhesions between the tentorium and cerebellar surface are sharply dissected using microsurgical techniques. Median bridging veins (vermian group) are divided if necessary, but paramedian and lateral bridging veins (hemispheric group) are preserved. Paramedian bridging veins are dissolved from the cerebellar surface for approximately 10 mm to obtain enough space between the tentorium and cerebellum (Figure 3). In the event that many median bridging veins are encountered on the vermis, we preserve these veins and divide a single or few paramedian bridging veins. Then the approach to the pineal region runs between the culmen and the cerebellar hemisphere (Figure 4). In both cases, we preserve the lateral bridging veins completely.

After supracerebellar dissection, the thick arachnoid membrane of the quadrigeminal cistern is sharply opened, and the withdrawal of cerebrospinal fluid (CSF) causes a reduction in tension of the cerebellum. If the patient undergoes the operation in the sitting position, retractors are usually not necessary, because gravity provides a sufficient space between the tentorium and cerebellum. Even with the patients in the Concorde position, we obtain the necessary surgical space only by mild and short-term cerebellar retraction, along with raising the upper part of patient’s body and withdrawing CSF. Continuous retraction of the cerebellum should be avoided to prevent cerebellar contusions and venous congestion caused by stretching or thrombosis of bridging veins and veins of the Galenic draining group.

The precentral cerebellar vein is now visible on the median surface of the tumor. The vein of Galen is detected above the precentral cerebellar vein, and bilateral basal veins of Rosenthal draining into the vein of Galen are also detected on the tumor along the tentorial incisura (Figure

5). The internal cerebral veins cannot be observed before resection of the tumor. The tectal segments of the bilateral medial posterior choroidal arteries feeding the tumor are visible near the tentorial edge.

Biopsy of the tumor should first be performed to determine the intraoperative pathological diagnosis with frozen sections of the tumor tissue. The tumor can be removed piecemeal beside the precentral cerebellar vein and under the basal veins of Rosenthal (Figure 5). We usually attempt to preserve the precentral cerebellar vein. However, if the precentral cerebellar vein blocks the surgical route, it should be divided as distally from the vein of Galen as possible, in the interest of preventing the progression of venous thrombosis to the vein of Galen (Figure 6A). If the thick superior vermian vein and the precentral cerebellar vein form the common trunk called "superior cerebellar vein," and if they interrupt the surgical route, the superior cerebellar vein should be divided at a point proximal to the junction of the superior vermian vein and precentral cerebellar vein, regarding both the prevention of the progression of venous thrombosis and the collateral venous circulation (Figure 6B). The basal veins of Rosenthal and the vein of Galen must be preserved completely.

The tumor is first decompressed by excavation using an ultrasonic aspirator, followed by dissection from the surrounding neural structures and veins. During the removal of dorso-rostral portion of the tumor, it is important to dissect and completely preserve the internal cerebral veins. More rostral portions of a tumor are less adhesive to surrounding structures because their bounding tissue is the ependyma of the third ventricle. However, it is sometimes difficult to dissect the ventro-caudal portion of the tumor from the quadrigeminal plate.

Neuronavigation

In general, neuronavigation can provide excellent planning of the craniotomy and approach and it was employed in all of our cases. Knowledge of the anatomy of the pineal region is absolutely irreplaceable, and neuronavigation can only assist slightly in deep lesions that progress into the thalamus. However, brainshift in the sitting position or even the Concorde position after CSF release is significant, and one should not rely heavily on preoperative imaging during intradural surgery in the pineal region.

Patients and surgical results

All 26 patients harboring a pineal-region tumor were treated microsurgically the through ITSC approach (23 in the sitting position and three in the Concorde position). Twenty-five patients had headaches and six patients initially suffered from seizures. The mean diameter of all of the tumors was 19 mm (7 to 43 mm). Before surgical resection, 25 patients underwent treatment for obstructive hydrocephalus. The patients received temporary ventricular drainage (EVD) (15 cases), permanent ventricular peritoneal (VP) shunt (four cases), or endoscopic third ventriculostomy (ETV) (six cases). The VP shunts were previously placed by the referring doctors/clinics. The choice placement of EVD or ETV depended on the experience of the surgeon on call during the initial presentation with hydrocephalus. Complete removal of the tumor was achieved in 23 of the 26 patients (88.5%), and subtotal removal was achieved in three (11.5%). Pathological diagnoses revealed pineocytomas in ten patients, astrocytomas in six patients, pineal cysts in four patients, and malignant gliomas in two patients, as well as one case each of ependymoma, pineoblastoma, anaplastic choroidolexus papilloma, and metastasis. Subtotal removal was performed in both malignant gliomas and the anaplastic choroido plexus papilloma. All five patients with malignant cases received adjuvant radiotherapy treatment.

The mean follow-up time of the surgically treated patients was 23.7 months (1 to 66 months). The mean hospital stay was 10.5 days. Clinical symptoms improved in 25 of 26 patients after the operation, and deterioration was observed in none of them after resection of the tumor. There was no mortality. Two patients experienced temporary symptoms after surgical resection: one presented Parinaud syndrome and the other displayed intermittent diplopia. However, both patients recovered completely. There were no cases with complications caused by injuries to the venous systems.

Discussion

The group of tumors in the pineal region is heterogeneous (27). Benign or low-grade tumors, such as pineocytomas, astrocytomas, ependymomas, meningiomas, and mature teratomas account for approximately one third of all pineal-region tumors (2, 6). Surgical resection is important in the treatment of these tumors. Pineal cysts are generally thought to result from the normal involution of the pineal gland and, incidentally, can be detected through radiological examinations (2, 16).

These cysts usually do not require surgical treatment. However, if mass effects, with compression of the aqueduct cerebri or quadrigeminal plate, are present, they may cause obstructive hydrocephalus, gaze paresis, or other neurological symptoms, then a surgical resection should be indicated.(28) For patients with malignant parenchymal or astrocytic tumors, such as pineoblastomas and glioblastomas, surgical resection plays an important role in ensuring the best starting conditions for adjuvant radiation and chemotherapy and in determining the tumor's histopathological etiology (1, 4, 14). Germinomas are highly sensitive to nonsurgical treatment and should be referred to radiation or chemotherapy (7). When the intraoperative pathological diagnosis of germinoma is determined, further surgical resection is no longer needed, and the patient should receive radiation or chemotherapy to treat the residual tumor and surrounding ventricle (7). The 20-year survival rate of patients with germinoma is expected to be more than 80% for those treated with radiation therapy with or without chemotherapy (8). For malignant germ-cell tumors, such as yolk-sac tumors, choriocarcinomas, and embryonal carcinomas, preoperative chemotherapy combined with radiation therapy and followed by tumor removal is reported to be highly effective (29). Therefore, if the preoperative levels of AFP and/or β-HCG in the serum or CSF strongly suggest a malignant germ-cell tumor, chemotherapy and radiation therapy may be indicated as the first treatment option, and surgical resection should be considered only for residual, therapy-refractory tumors.

Although surgical resection plays a significant role for many kinds of pineal-region tumors, the deep location of this region and the existence of surrounding vital veins belonging to the Galenic draining group make the surgery challenging for neurosurgeons. Bruce and Stein reported a large series of surgeries for pineal-region tumors (1). They performed 160 operations for pineal-region tumors, mainly using the ITSC approach (86%). The operative mortality was 4%, with 3% permanent major morbidity. A gross total tumor removal was possible in 31 of 107 malignant tumors. Konovalov et al. also reported a large series of surgeries for histologically verified pineal-region tumors (3). In all, 255 tumor-removal surgical procedures were performed in 244 patients. The OTT approach was used in 138 procedures (54%), and ITSC was used in 87 cases (34%). A total tumor removal was achieved in 148 operations (58%), a subtotal removal was achieved in 74 cases (29%), and a partial tumor removal was achieved in 33 operations (13%).

In our serial, we present preferable results obtained by microsurgically resecting tumors of the pineal region via the ITSC approach. One of the most crucial points is preserving the venous flow of the Galenic draining group. It is essential to preserve completely the bilateral basal veins of Rosenthal, the bilateral internal cerebral veins, and the vein of Galen to avoid serious surgical complications, such as venous infarction in the thalamus, hypothalamus, or midbrain. The ITSC approach is more suitable for protecting veins of the Galenic draining group compared with the OTT approach because surgeons are able to access the tumor from the spaces under the bilateral basal veins of Rosenthal, where no other important veins exist beyond the precentral cerebellar vein. Using the sitting position, gravity reduces the venous pressure, which makes the dissection of veins from the tumor easier. It has been generally reported that the precentral cerebellar vein can be sacrificed without any clinical symptoms (18, 22, 30). However, Kanno reported a case with thrombosis of the basal veins of Rosenthal and internal cerebral veins, which were initiated by division of the precentral cerebellar vein and followed by fatal hemorrhagic infarction (31). In this case, the precentral cerebellar vein was coagulated too close to the confluence of the basal veins of Rosenthal. Kanno concluded that the precentral cerebellar vein should be coagulated at the peripheral point as far from the confluence as possible when it must be divided. We always attempt to preserve the precentral cerebellar vein. If it must be sacrificed, we divide it with regard to the prevention of the progression of venous thrombosis and collateral venous circulation.

We consider that the preservation of bridging veins between the cerebellar surface and the tentorium is also an important point for the successful resection of pineal-region tumors. As bridging veins and superficial veins of the cerebellum have frequent communications with one another, some of them can be divided without any adverse effects (21, 26). On the other hand, Page reported that severe cerebellar swelling occurred after the division of all vermian bridging veins and one hemispheric bridging vein. He strongly advocated the importance of preserving the hemispheric bridging veins (12). We always divide only the thin bridging veins and try to preserve the thick ones. If some of the thick bridging veins strongly restrict the surgical space, we divide only median bridging veins and dissect paramedian veins for several millimeters from the cerebellar surface to gain enough space between the tentorium and the cerebellum. We preserve hemispheric (paramedian and lateral) bridging veins completely, out of consideration for their collateral circulation. When median bridging veins are prominent and restrict the surgical space, we divide a few of the paramedian bridging veins. In those cases, we approach the pineal region through the space between the culmen and cerebellar hemisphere and preserve the median and lateral bridging veins.

We always attempt to minimize the retraction of the cerebellum during surgery. Kanno reported that strong retraction of the cerebellar vermis worsened the auditory brain stem response; therefore, they usually did not use the retractor in the course of surgery using the ITSC approach (31). Bruce and Stein reported that brain retraction and sacrifice of bridging veins occasionally cause complications after surgeries on pineal-region tumors using the supratentorial approach (1). Prolonged retraction of the brain provokes local congestion by compression of the cortical venous network, causes a reduction in the venous flow by stretching the bridging vein, and can cause thrombosis of veins (30).

Conclusion

We present a microsurgical technique for the resection of pineal-region tumors using the ITSC approach with special regard to the venous aspect. We conclude that i) preserving the venous flow of the Galenic draining group, ii) preserving the thick bridging veins of the tentorial surface of the cerebellum, especially the hemispheric bridging veins, in consideration of the collateral circulation, and iii) minimizing retraction of the cerebellum during surgery to avoid adverse effects, both due to direct cerebellar compression and due to disturbance of venous circulation, are the most important considerations for achieving a successful operation and patient outcome in tumors of the pineal region.

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Figure Legend

<u>Figure 1:</u> Infratentorial supracerebellar view of the pineal region of a human cadaver. Veins belonging to the Galenic draining group are prominent in the pineal region. From the posterior side, the precentral cerebellar vein (Prec. Cereb. V.), internal occipital veins (Int. Occip. V.), basal veins of Rosenthal (Basal V.), and internal cerebral veins (Int. Cerebr. V.) drain into the vein of Galen (V. Galen). The superior vermian veins (Sup. Verm. V.) drain into the precentral cerebellar vein, and the pineal vein (Pineal V.) drains into the internal cerebral vein. The pineal body (Pineal) is supplied from the branches of the medial posterior choroidal artery (Med. Post. Chor. A.), and the P3 segments of the posterior cerebral arteries (P3) run around the lateral parts of the pineal region. Tent. Incis. = tentorial incisura, Sup. Coll. = superior colliculus, Inf. Coll. = inferior colliculus.

<u>Figure 2:</u> Indication of skin incision, craniotomy, and dural incision. A midline linear skin incision is placed from approximately 4 cm above the inion (arrow) to the level of the spinous process of the axis (red line). A bilateral suboccipital craniotomy is performed with its upper line placed above the superior margin of the transverse sinus (blue line). The dura mater is incised transversely 5 to 10 mm below the inferior margin of the transverse sinus (red dotted line).

<u>Figure 3:</u> Overview of veins involved with using the ITSC to gain access to the space between the tentorium and the cerebellum; thin bridging veins and only median thick bridging veins can be

coagulated and divided (red lines), and paramedian bridging veins are preserved while being dissected for approximately 10 mm from the cerebellar surface with microscissors and dissectors (red dotted lines). By intermittently retracting the culmen or medial tentorial surfaces of the cerebellar hemispheres with a suction tube, surgeons can reach the posterior tentorial incisura (arrows) and the pineal region.

Figure 4: When median bridging veins are prominent and paramedian ones are thinner and smaller in number than median ones, a few paramedian bridging veins may be divided (red line), and almost median ones are preserved by dissecting some of them from the cerebellar surface (red dotted line). Surgeons can approach the posterior tentorial incisura (arrow) and the pineal region through the space between the culmen and cerebellar hemisphere.

Figure 5: Tumor removal through the space beside the precentral cerebellar vein (Prec. Cereb. V.), under the basal vein of Rosenthal (Basal V.) and the vein of Galen (V. Galen). Tent. Incis. = tentorial incisura.

Figure 6:

A: If necessary, the precentral cerebellar vein (PCCV) can be divided as distal from the vein of Galen (VG) as possible, together with the thin superior vermian veins (SVV) (red lines), to prevent the progression of venous thrombosis to the vein of Galen.

B: The superior cerebellar vein (SCV) can also be divided at a point proximal to the junction of the superior vermian vein and the precentral cerebellar vein (red line), to prevent both the progression of venous thrombosis and the collateral venous circulation.

Figure 1:

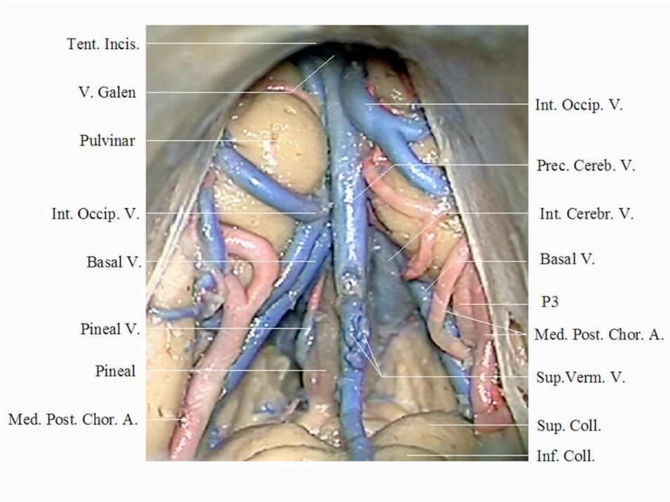

Figure 2:

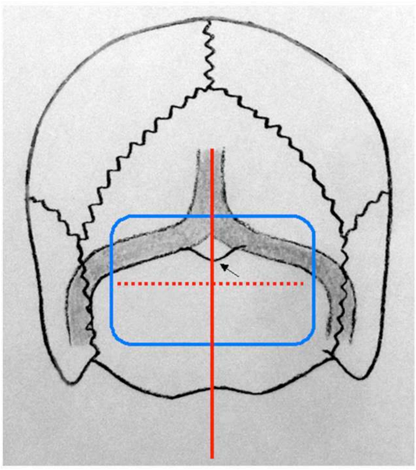

Figure 3:

A

B

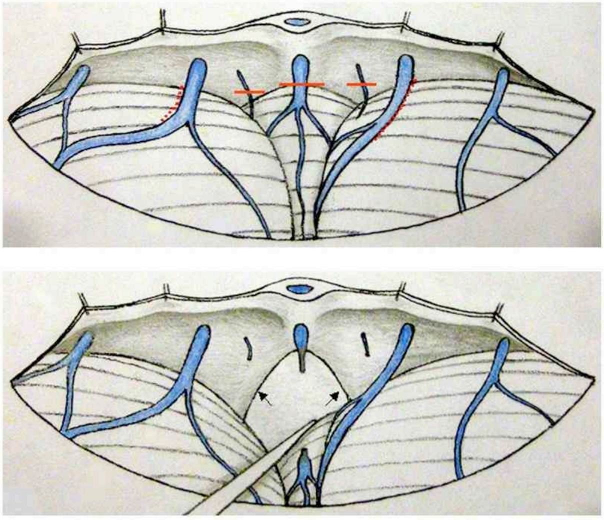

Figure 4:

A

B

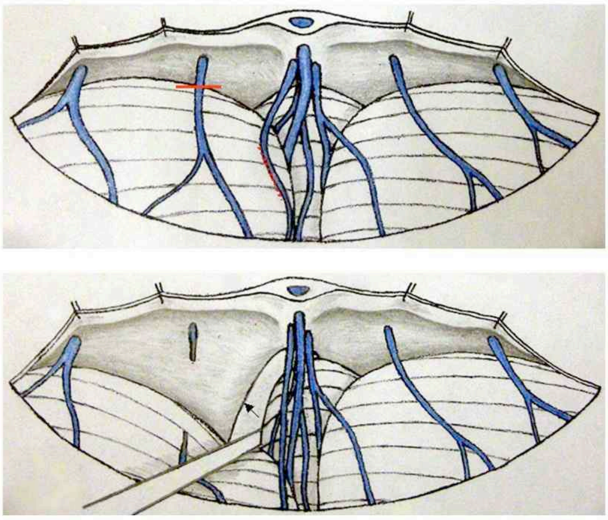

Figure 5:

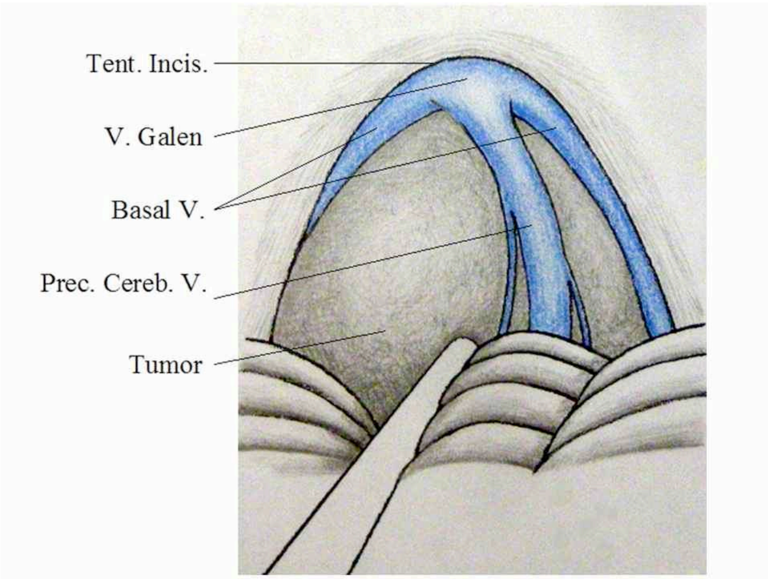

Figure 6: